DOI:
10.1093/bjsw/bcy116

Document Version
Peer reviewed version

The role of social work in supporting people affected by Creutzfeldt-Jakob Disease (CJD): a scoping review

Jill Manthorpe and Peter Simcock

Abstract

Social work practice in England and the wider United Kingdom is curiously under-informed about its own practice in supporting people affected by Creutzfeldt-Jakob Disease (CJD). This paper reports the findings of a scoping review undertaken in 2018 of what is known about social work support for people with CJD and their families. Conceptual and thematic analysis of the limited literature identified in the review enabled us to develop a consensus on reported themes: rapid decline and the need for quick responses; families' need for support; and the use of case studies and case reports. The review identified significant gaps in the evidence, particularly as the literature currently published pre-dates the contemporary legal framework for social work practice in England. We therefore argue that there is clear scope for further research, investigating the specific social work role in supporting people affected by CJD, potentially providing the profession with greater confidence about its potential contribution, and evidence of its potential and actual effectiveness.

Keywords:

Creutzfeldt-Jakob Disease; CJD; social work; dementia; scoping review

Introduction

"There was a social worker, but all she provided was incontinence pads." This statement was reported in the national press (Arthur 1998) as being made in a father's submission to the Philips BSE (bovine spongiform encephalopathy) Inquiry (Philips *et al.* 2000) about the care of his daughter who died from variant Creutzfeldt-Jakob Disease (vCJD). It potentially illustrates social workers' lack of awareness, knowledge and understanding of the condition, identified by a CJD Support Network Survey (Biggs 2003) at the time of the emergence of BSE/vCJD. It also reflects the reality that whilst improvement in social care services for people with CJD has been reported (Body and Glasson 2005), work with people who are affected by the condition remains outside the experience of most social workers, owing to its rarity.

This paper reports the findings of a scoping review of what is known from the research literature about social work support for people with CJD and their families. The review formed part of background research for the development of new Guidelines for Social Workers in England on CJD, published in 2018 (Simcock, Manthorpe, Tesfamichael and Mead, 2018) by the English Department

of Health and Social Care and commissioned by its Chief Social Worker. These Guidelines revise those produced in 1998 (and subsequently amended and re-published in 2003) by a social worker, Derek Biggs (Biggs 2003) of Cambridgeshire Social Services. The need for new Guidelines was evident following the advent of the Mental Capacity Act 2005, and the Care Act 2014, which consolidated previous legislation and added new responsibilities to local authorities in England. Furthermore, since the first Guidelines, there is now greater emphasis on the potential for social workers to help support people with dementia and their carers. As the UK Alzheimer's Society (2015) notes, CJD is one of the rarer causes of dementia and there is increasing interest in responding to the different forms of dementia and not seeing its experiences as being homogeneous. Overall, within wider health services, a recent Cochrane Review on the notification and support for people exposed to the risk of CJD commissioned in Australia (Ryan *et al.* 2011) noted that:

> Effective ways to inform and support people at risk over time are needed in relation to infection control measures and to promote people's capacity to manage their health care, as well as to promote healthcare responses that are appropriate to the person's individual risk level.

Such conclusions are also relevant to social work, as the profession seeks to promote individuals' well-being, undertake empowering risk assessments, and tailor individual and personalised care and support.

Creutzfeldt-Jakob Disease (CJD): Defining the condition

CJD is a degenerative, always fatal disease caused by abnormal forms of naturally occurring protein found in the brain (known as prions). Average onset and duration of the disease vary by type, though rapid deterioration and progression to death, often within the year, is usual in the most common type (Barnett and McLean 2005, Freeman *et al.* 2010). CJD is however rare; according to Mackenzie and Will (2017) in developed countries mortality rates of 1.5–2 cases per million are the best estimates on current evidence.

Clinical discussion of CJD often refers to three main types; sporadic or classical CJD being the most common form, accounting for about 80-90% of all cases (Rentz 2008, Ryan *et al.* 2011). What triggers sporadic CJD is unknown, but it is not thought to be inherited or otherwise transmitted interpersonally. A condition mainly affecting those in late middle age or older age, sporadic CJD results in a rapidly progressive dementia (Barnett and McLean 2005, Das *et al.* 2012). Second, are very rare cases of genetic or inherited CJD, which usually have slower rates of progression after symptom onset (Barnett and McLean 2005). Third, are cases of acquired or iatrogenic CJD, caused by exposure to

prion disease during medical treatment (first case in 1974) or from the diet. This third type includes variant CJD (initially termed new variant CJD), caused by dietary exposure to Bovine Spongiform Encephalopathy (BSE), a prion disease found in cattle. Variant CJD (vCJD) was widely publicized in the UK media and seen as a matter of public policy (as summarised in Table One supplementary document). It affects younger people than sporadic CJD and has a longer duration period, approximately 14 months. Initial symptoms include psychiatric or mental health problems before the onset of neurological difficulties (Barnett and McLean 2005).

Diagnosis of all forms of CJD can be lengthy and difficult (Barnett and McLean 2005); not simply because it is rare, but also because some symptoms might suggest a dementia or another form of neurological disease, or mental health conditions such as depression (Barnett and McLean 2005). Initial symptoms of visual impairment have also been noted (Freeman *et al.* 2010). European Centre for Disease Prevention and Control (2017) figures report that from 1996 - 2013, 229 cases of vCJD have been identified from 11 countries: 177 from the United Kingdom (UK), 27 from France, 4 from Ireland, 4 from the United States, 5 from Spain, 3 in the Netherlands, 2 each from Portugal, Italy and Canada, and 1 each from Japan, Taiwan and Saudi Arabia. Over the same period, an estimated 2,000 cases of sporadic CJD were identified in the UK. Approximately 100-130 people are newly diagnosed with CJD every year in the UK (Authors 'Own). Thus for social workers in the UK, while the condition is not often encountered, when all types of CJD are taken into account, it is likely that there will be cases locally, albeit rare in comparison to other neurological problems including Alzheimer's type dementia. Social workers in services for older people may need to respond to those with sporadic CJD. However, those in services for working age adults may also receive referrals for those with vCJD, and as the House of Commons Science and Technology Committee (2014:47) report '*After the Storm'* notes:

> Variant Creutzfeldt-Jakob Disease (vCJD) is not like other infectious diseases. Caused by a mysterious pathogen, which we are still only just beginning to understand, vCJD is an invariably fatal disease of sudden onset, which has historically inflicted on its young victims a progressive dementia more often seen in the oldest and sickest members of society.

Insert Table One Supplementary Document Here

Method

The aim of this review was to explore what is known about social work practice with people who have CJD and their families. It was undertaken to inform new guidelines for social workers in England on CJD but also to contribute to the evidence base for social work practice with people with dementia and their

carers. Our initial searches revealed the limited published research on the subject and suggested that we should adopt a scoping methodology for our review. Scoping reviews (Arksey and O'Malley 2005) are increasingly popular methods to map the body of literature on a given topic in a timely, transparent and rigorous way (Pham *et al.* 2014; Moriarty *et al.* 2017). Furthermore, as Peterson *et al.* (2017:14) note:

> [a] scoping review can provide a broad overview of the evidence and controversies before a clinical practice guideline is available [and] serve as one source of information in the development of practice guidelines.

Moriarty *et al.* (2017) observe that in social care research they are currently being primarily used for 'reconnaissance' purposes (Peters *et al.* 2015) to provide an overview of a potentially large and diverse body of literature on a broad topic. However, scoping reviews can also be useful when there is very little material on a topic and where material needs to be interrogated to see if the topic may be 'hidden' within findings or discussion. Whatever the range of material, a scoping review's findings can help inform new primary research or indicate whether a systematic review is feasible.

While increasing in popularity, the conduct of scoping reviews varies considerably and there are calls for more debate about the ways in which they are undertaken and reported in order to achieve greater transparency and consistency (Peters *et al*. 2015, Tricco *et al*. 2016). We followed the three-step process recommended for scoping reviews (Peters *et al*. 2015) by systematically searching for primary research, literature reviews and other types of resource, such as toolkits or guidance concerned with social work practice with people affected by CJD, published from 2003 up to December 2017. First, we identified relevant bibliographic databases that covered research such as PubMed, Cumulative Index to Nursing and Allied Health Literature, ASSIA, and Social Care Online.

Two sets of terms were sought. Set one included Creutzfeldt-Jakob Disease (CJD) (any type) and prion disease. Set two included social work and social care. Next we searched the Internet to identify grey literature not formally published in academic journals or books to identify government reports and guidance, and other relevant material that may not have been abstracted in bibliographic databases. This included searches of the website of the Social Care Institute for Excellence and the Alzheimer's Society. Table Two (supplementary document) presents the search terms adopted and a summary of the inclusion and exclusion criteria for the review. Table Three (supplementary document) summarises the results from the searches.

Insert Tables Two supplementary document

and Table Three supplementary document Here

We undertook conceptual and thematic analyses to summarise the review's findings. First, both authors read all the material, made notes, and developed a table of key findings and other relevant details. We then discussed the findings to reach a consensus on the themes reported. While a small minority of scoping reviews include elements of quality appraisal (Peters *et al.* 2015, Pham *et al.* 2014), the material accessed did not lend itself to this scrutiny. However, information on the design and scope of the included studies is recorded in Table One (main document)

Insert Table One (main document) Here

Findings

Limited Mention of Social Work Practice

The published literature on CJD in which social work is mentioned is small and a high proportion appears to mention a social worker only in passing. Some material from outside the UK has been included where the social work role was identifiable but there is similarly little information available internationally. A small part of the literature speaks of inter-governmental collaboration (Gerodimos 2004) which, while not mentioning social work specifically, presents some background context of the political debates about culpability for the BSE 'crisis', cited as being 'one of the worst policy disasters experienced by a UK government in recent years' (Beck *et al.* 2007:2). Authors such as Body and Glasson (2005) chart the development of Department of Health responses in England and recent legal judgments of the time, and note some improvement in NHS and local authority care and support, with the additions of payments and compensation from the vCJD Trust. Specifically they report that the national CJD care team is able to help with the cost of counselling and psychiatric support if only private sector counselling is available. However, in their descriptive overview of the work of the national CJD care team and outline of the use of national CJD care package funds, Barnett and McLean (2005) observe limited demand for such private counselling. They suggest that this may indicate that people are unaware that such assistance is available. At the local level, Barnett and McLean (2005) observe that social workers may be members of the multi-disciplinary team and act as key workers; however, there is no exploration of social work practice in this context.

Rapid Decline and the Need for Quick Responses

Much of the literature relates to the early days of recognition of CJD and in particular the emergence of vCJD. As de Vries (2006) argues in her thesis,

professional responses to this new condition need to be developed overall through accumulation of experience of the rapid decline of patients and developing knowledge of the problems facing both patients and their family members. The rapidly progressive nature of CJD is also highlighted elsewhere in the literature (Freeman *et al*. 2010, Rentz 2008), and the earlier good practice guidelines repeatedly emphasise the need for social workers to respond without delay (Biggs 2003). Writing in the United States (US) context, Rentz (2008) observes that time is 'of the essence' at diagnosis to make arrangements and tailor support as symptoms change. She also suggests that people with CJD and their families may not have time to explore care options because of the rapid onset of the condition. Relevant to the UK context is her suggestion therefore that executing advance directives (in England and Wales these would be advance decisions) may need to be considered if possible.

Families' Need for Support

The need for family support, in addition to that for the person with CJD, is noted across the literature (Das *et al*. 2012, Morris 2010, Rentz 2008, Wickenden 2006). De Vries' (2006) thesis contains observations from hospice staff, including a social worker, and from family members, with social work quotations used to illustrate the reactions of family members to the distressing situations of their relatives and accounts of their engagement with other services. In her thesis she proposes ways in which hospice staff may learn more about patients' clinical needs but also about communications with patients' relatives. The context of this study was that of the early days, when BSE was being covered in the media (start of this millennium) and de Vries reports accounts of some patients' children being bullied arising from the 'mad cow' connotations or stigma of BSE. In respect of care packages being arranged at home, she reports social work acknowledgement of some families' great efforts to sustain these and seeming antipathy to professionals visiting the home.

Some of de Vries' work prior to her thesis is presented in an earlier publication (de Vries *et al*. 2003), which is described as an inductive qualitative study (a case study) of a woman with vCJD receiving palliative care in a hospice setting. This is described as a pilot study for a larger research project (the thesis). While noting the presence of the patient's family when visiting their relative in the hospice, there is no mention of the patient's children or social work, and no mention of hospital social work or palliative care social workers, albeit that the authors acknowledge that providing care for people with vCJD is a stressful experience at all stages. In a further development from her thesis, (de Vries and McChrystal 2010) argue that attachment theory is a valuable lens through which to consider the distress of some patients with dementia who may feel abandoned in care settings and are expressing this loss through distress.

In the US context, Freeman *et al.* (2010), drawing on two case studies, similarly observe a reliance on family carers, though they note that a move to a long-term care facility may be necessary. Again from the US context, (Rentz 2008) also considers the needs of family members, noting that they may need support into the bereavement period; feelings of loss may be unresolved during intensive caregiving and the rapid progression of the disease. She notes the emotional pain of both patient and family and their need for emotional support. It is therefore unsurprising that Freeman *et al.* (2010) recommend that compassionate care should be prioritised. More controversially perhaps, they also raise the question of whether it is always right to inform the patient of their diagnosis. While Wickenden (2006) advises clinicians is to tell family the 'hard facts', such as CJD being a terminal condition, even though diagnosis uncertainty is emphasised, Rentz (2003), in an earlier paper, acknowledges that the communication of diagnosis may be distressing for all concerned.

Case Reports and Case Studies

Case reports from medical and nursing practitioners are common features of the CJD literature that focuses on care and thereby mentions social work (although de Vries (2006) is the only one to mention accessing social work records during her research). This case study approach was also undertaken by Wickenden (2006) a psychiatrist involved in the assessment of two patients presenting to British military health services. The first case he discusses is that of an unmarried 25 year old man; the second a 32 year old married father. Such cases are used to illustrate the difficulties of diagnosis and the need for multiple assessments including, in his accounts, consideration of the possibility that the symptoms might be imaginary. In this paper there is no discussion of social work despite parents and spouse and even the men's young children facing what are described as irritable and harsh responses from each patient. This paper is however one of the few to mention the national CJD support initiatives that were developing at the time.

While Barnett and McLean (2005) offer a brief overview of the National CJD Care Team, Morris (2010) provides further detail of the counselling service for CJD patients in the UK National Prion Clinic, that has provided information, advocacy and support for patients and families since 1997. In this paper, some case examples are provided from the Clinic's practice; specifically there is mention of one patient who felt 'betrayed by her social worker' and opted not to attend a case conference. Another who feared sexual abuse is discussed; another with perceptual disorders affecting recognition. Morris reports that family meetings and case conferences can help co-ordinate care and that talking to other professionals is more useful than sending a referral letter. One example of a

patient whose (social care) services were being unreliable is provided. Also mentioned is the presence of funding disputes between the local authority and the NHS, to the extent that social services staff were managing complaints rather than meeting patient and family needs. Another case account features more positive inter-agency working with a patient's family, including a child. A further case example concerns a young person with the familial or inherited form of CJD. Here the Clinic's development of a care package for the parent was of immediate concern, but Morris notes that this young person would need continued support as inherited CJD may not develop so rapidly as other forms of the condition (Barnett and McLean 2005). Morris, employed in the National Prion Clinic at the time of the article, argues that professionals also need support in the form of reflection, collaboration, and discussion to relieve their stress.

Naturally, the fewer the cases discussed the more detail can be provided in case study research. Das *et al.* (2012) focus on two cases of people with Sporadic CJD from their psychiatric practice in Southern England. In the first, a 70-year-old woman is described as needing 'home nursing care' but later moving to a hospice prior to death. In the second case of a 69-year-old man whose condition also deteriorated rapidly, he is described as receiving services at home including liaison psychiatry and palliative care; and then dying in hospital. Overall, the authors note that many of the patients they encounter have both mental and physical symptoms, sometimes making it hard to find suitable services for them, and that CJD's unusual presentations mean patients may fall 'between stools' of services and that families get 'short shrift'. As with other writers from the UK, they mention the National CJD Surveillance Team as providing advice to local services and families.

A final document reviewed is that of the 2003 amended version of the *Good Practice Guidelines for Social Services Professionals* produced by social worker Derek Biggs (Biggs 2003). Originally produced in 1998, these were later complemented by the Department of Health (2000a) *CJD: Guidance for Healthcare Workers*. Biggs (2003) reports that these guidelines were stimulated by concerns from the CJD Family Support Network, whose member survey had revealed not only variable responses but also reports that social services are: 1) too little and too late; 2) uninformed about CJD; 3) feel isolated and uncertain of their practice; and 4) lack co-ordination. The Guidance advises the need for a key worker to be appointed, timely action, offers of carer support, and collaboration between agencies. Some information is provided on the then new funds for patients and carers, with specific note (para. §6.11) that people with any form of CJD should be exempt from social care charging, reflecting the '*Fairer Charging Polices*' at the time (Department of Health 2013). Other feedback from professionals, reported in the first Guidelines, had suggested that social services need to transcend just care management to include social work casework and

counselling. This reflects Morris' (2010) later assertion about the effectiveness of emotional and talking support.

Discussion

The literature reviewed suggests that social work practice with people affected by CJD has little evidence on which to develop or upon which to assess the quality of its professional response. That the condition is rare is likely to be a plausible explanation for this. Furthermore, CJD may not be well known among younger age groups that are not aware of the political crisis surrounding BSE and the major media spotlight on the condition in the 1990s. The evidence does suggest that while other professionals offer some illustrative case studies that can be used with their peers or in professional development or training, this is not something that has been seen as part of the professional culture of social work. While there are ongoing concerns about risks, these have not prompted interest in what social work can offer to those affected and to wider professional teams. This resembles some other conditions that are not common, such as Huntingdon's disease (see, for exception, Mantell 2010). While there is interest in social work support for people with dementia of the Alzheimer's disease type, this follows a framework of 'living well with dementia' (Department of Health 2000b) and promoting the advantages of early recognition and thereby possible lessening of symptoms. Social work practice appears to be disadvantaged by the lack of opportunities to discuss cases through refection on published case studies in the same way as other professionals are able to build up their own practice evidence. However, the case study approach that is evident from this review should not be seen as providing multiple sources of data about 'the case' facilitating detailed analysis (see Harrison *et al.* 2017) but is more descriptive and from a clinical or scientific perspective.

In only one doctoral study were social workers interviewed about their perspectives; indeed this study was also interested in accessing social work records although this was not reported in any depth in the thesis (de Vries 2006). This may be because several studies focus on clinical services, both those at the time of diagnosis and also palliative care, as well as being conducted by a health professional member of the clinical team. The inclusion of social workers in this thesis did not reveal unique insights but did offer another perspective from those who were not undertaking clinical roles in the hospice. This includes the importance of offering support not only to the person with CJD, but also their families, a point acknowledged across the literature reviewed. There may be social work practice interventions to hear about in respect of family support, bereavement services and reducing possible family conflict. While other researchers have addressed their own professional concerns, there remain opportunities to consider working inter-professionally at local levels and also with centres of national support.

Scoping reviews offer the opportunity to identify where the gaps in the evidence may lie

and social work with people affected by CJD is an area where there are many gaps. The current legal context in England of the Care Act 2014 and Mental Capacity Act 2005 post-date the studies included in this review and nothing is known about how personal budgets are impacting upon those affected by CJD and their families, to their benefit or otherwise. The new Guidelines for Social Workers in England (Simcock *et al.* 2018) offer the profession an opportunity to consider how it may build up evidence of practice following the implementation of the Care Act 2014 in particular. Moriarty and Manthorpe (2016) note that there is considerable satisfaction with palliative care social workers among service users and carers but evidence of unmet demand. Such a view is not informed by the experiences of people affected by CJD who may additionally have contact with a range of social workers or find access difficult. We do not know if social workers are taking on the role of case co-ordination for individuals locally and what such practice involves, albeit that the Department of Health (2000a) asserts that the important role of key worker when coordinating care for those with CJD can be undertaken by social workers.

The vexed question of funding sources may also be problematic for people affected by CJD in England, particularly where disagreement between agencies as to financial responsibility results in delay in care and support provision. Operation of the 'rationing device' of NHS Continuing Healthcare (a care package provided in any setting to those with a primary health need and fully funded by the NHS) is an area where social workers get involved. As Parliament's Public Accounts Committee (2018) acknowledges, this is a hugely complicated system, even when the Fast Track Pathway Tool for people with rapidly deteriorating conditions that may be entering a terminal phase, such as CJD, is followed. While the recently updated National Framework for NHS Continuing Healthcare (Department of Health and Social Care 2018), coming into operation in October 2018, has offered increased clarity in relation to some processes, commentators maintain that key concepts, such as 'primary health need', remain unclear (Parsons 2018, Schwehr 2018). Furthermore, while the *Fairer Charging Guidance* (Department of Health 2013:para. 75) automatically exempted people with *any form* of CJD from social care charges, it appears that the *Care and Support (Charging and Assessment of Resources) Regulations 2014* restrict this exemption to those with *variant* CJD only (section 4).

Conclusion

Social work practice in England and the wider UK is curiously under-informed about its own practice in supporting people affected by CJD, which may be surprising internationally since the UK has experience of this work in far greater numbers than other countries. The published literature on CJD in which social work is even mentioned is small and a high proportion of this appears to mention a social worker only in passing. However, conceptual and thematic analysis of the limited literature identified in this scoping review, enabled the authors to develop a consensus on

reported themes: rapid decline and the need for quick responses; families' need for support; and the use of case studies and case reports. While the scoping review supported the development of new guidelines for social workers in England, it also identified significant gaps in the evidence, particularly as the literature currently published pre-dates the contemporary legal framework for social work practice. There is therefore scope for further research, exploring the social work role in supporting people affected by CJD. There may also be scope to investigate the social work role when supporting other rare forms of dementia and neurological problems and potentially providing the profession with greater confidence about its potential contribution, and evidence of its potential and actual effectiveness.

Acknowledgements and disclaimer

To be inserted following review.

References (* indicates inclusion in the review)

Alzheimer's Society (2015) 'Rarer Causes of Dementia Factsheet', [online], available: https://http://www.alzheimers.org.uk/sites/default/files/pdf/factsheet_rarer_causes_of_dementia.pdf [accessed 19/06/2018].

Arksey, H. and O'Malley, L. (2005) 'Scoping studies: towards a methodological framework', *International Journal of Social Research Methodology,* 8(1), 19-32.

Arthur, C. (1998) 'CJD patients were left to suffer', *The Independent* [online], available: https://http://www.independent.co.uk/news/cjd-patients-were-left-to-suffer-1180911.html [accessed 20/06/2018].

*Barnett, F. and McLean, G. (2005) 'Care management of Creutzfeldt-Jakob Disease within the United Kingdom', *Journal of Nursing Management,* 13(2), 111-118.

Beck, M., Kewell, B. and Asenova, D. (2007) 'BSE crisis and food safety regulation: a comparison of the UK and Germany. Working paper number 38.', [online], available: http://eprints.whiterose.ac.uk/3477/1/beckm32007.pdf [accessed 20/06/2018].

*Biggs, D. A. (2003) *Creutzfeld-Jakob Disease: Good Practice Guidelines for Social Services Professionals (Amended Version),* Cambridge: CJD Support Network / Association of Directors of Social Services.

*Body, D. and Glasson, J. (2005) 'BSE and the particular British problem of variant CJD', *Clinical Risk,* 11(1), 14-24.

*Das, K., Davis, R., Dutoit, B. and Parsons, B. (2012) 'Sporadic Creutzfeldt-Jakob disease: a description of two cases', *International Psychogeriatrics,* 24(7), 1183-5.

*de Vries, K. (2006) *Caring for the person with variant Creutzfeldt-Jakob disease within the hospice service*, unpublished thesis University of Surrey (United Kingdom).

*de Vries, K. and McChrystal, J. (2010) 'Using attachment theory to improve the care of people with dementia', *International Journal of Work Organisation and Emotion,* 3(3), 287-301.

*de Vries, K., Sque, M., Bryan, K. and Abu-Saad, H. (2003) 'Variant Creutzfeldt-Jakob disease: need for mental health and palliative care team collaboration', *International Journal of Palliative Nursing,* 9(12), 512-20.

Department of Health (2000a) *Creutzfeldt-Jakob Disease: Guidance for Healthcare Workers,* London: Department of Health.

Department of Health (2000b) *Living well with dementia: a National Dementia Strategy,* Department of Health [online], available: https://http://www.gov.uk/government/publications/living-well-with-dementia-a-national-dementia-strategy [accessed 20/06/2018].

Department of Health (2013) *Fairer Charging Policies for Home Care and other non-residential Social Services: Guidance for Councils with Social Services Responsibilities,* London: Department of Health.

Department of Health and Social Care (2018) *National Framework for NHS Continuing Healthcare and NHS-funded Nursing Care,* London: Department of Health and Social Care.

European Centre for Disease Prevention and Control (2017) 'Facts about variant Creutzfeld-Jakob disease', [online], available: https://ecdc.europa.eu/en/vcjd/facts [accessed 19/06/2018].

*Freeman, J. W., Free, T., Shinozaki, E. and Koch, M. (2010) 'The sharp point of care: quality, safety and Creutzfeldt-Jakob disease', *South Dakota Medicine,* 63(8), 281-283.

*Gerodimos, R. (2004) 'The UK BSE crisis as a failure of government', *Public Administration,* 82(4), 911-929.

Harrison, H., Birks, M., Franklin, R. and Mills, J. (2017) 'Case Study Research: Foundations and Methodological Orientations', *Forum Qualitative Sozialforschung/Forum: Qualitative Social Research*, 18(1), Art. 19, available: http://www.qualitative-research.net/index.php/fqs/article/view/2655/4079 [accessed 19/06/2018].

House of Commons Science and Technology Committee (2014) *After the Storm? UK blood safety and the risk of variant Creutzfeldt-Jakob Disease Second Report of Session 2014-15,* House of Commons [online], available: https://publications.parliament.uk/pa/cm201415/cmselect/cmsctech/327/327.pdf (accessed 10/11/2018)

Mackenzie, G. and Will, R. (2017) 'Creutzfeldt-Jakob disease: recent developments', *F1000Research,* 6(F1000Faculty Rev), 2053.

Mantell, A. (2010) 'Under a cloud: carers' experiences of Huntington's disease', *Social Care and Neurodisability,* 1(2), 33-42.

Moriarty, J., Lipman, V., Norrie, C., Elaswarapu, R. and Manthorpe, J. (2017) 'Handovers in care homes for older people–their type, timing and usefulness. Findings from a scoping review', *Ageing & Society*, 1-21.

https://doi.org/10.1017/S0144686X17001301Published online: 27 November 201

Moriarty, J. and Manthorpe, J. (2016) *The Effectiveness of Social Work with Adults,* London, King's College London, https://www.kcl.ac.uk/sspp/policy-institute/scwru/pubs/2016/reports/Moriarty-&-Manthorpe-2016-Effectiveness-of-social-work-with-adults.pdf (accessed 10/11/2018)

*Morris, C. (2010) 'Counselling people with Creutzfeldt-Jakob Disease (CJD) and their families', *Dementia,* 9(4), 569-576.

Parsons, M. (2018) 'What's changed? Our commentary on the 2018 update to the National Framework', [online], available: http://www.beaconchc.co.uk/our-commentary-on-key-updates-to-the-nhs-chc-framework/ [accessed 05/07/2018].

Peters, M. D., Godfrey, C. M., Khalil, H., McInerney, P., Parker, D. and Soares, C. B. (2015) 'Guidance for conducting systematic scoping reviews', *International journal of evidence-based healthcare,* 13(3), 141-146.

Peterson, J., Pearce, P. F., Ferguson, L. A. and Langford, C. A. (2017) 'Understanding scoping reviews: Definition, purpose, and process', *Journal of the American Association of Nurse Practitioners,* 29(1), 12-16.

Pham, M. T., Rajić, A., Greig, J. D., Sargeant, J. M., Papadopoulos, A. and McEwen, S. A. (2014) 'A scoping review of scoping reviews: advancing the approach and enhancing the consistency', *Research synthesis methods,* 5(4), 371-385.

Philips, N., Bridgeman, J. and Ferguson-Smith, M. (2000) *The BSE Inquiry: Report, evidence and supporting papers of the Inquiry into the emergence and identification of Bovine Spongiform Encephalopathy (BSE) and variant Creutzfeldt–Jakob Disease (vCJD) and the action taken in response to it up to 20 March 1996),* London: The Stationery Office.

Public Accounts Committee (2018) *Continuing Health Care Funding,* Parliament [online], available: https://publications.parliament.uk/pa/cm201719/cmselect/cmpubacc/455/45502.htm [accessed 20/06/2018].

Rentz, C. A. (2003) 'Creutzfeldt-Jakob disease: two case studies', *American Journal of Alzheimer's Disease and Other Dementias,* 18(3), 171-180.

*Rentz, C. A. (2008) 'Nursing care of the person with sporadic Creutzfeldt-Jakob disease', *Journal of Hospice and Palliative Nursing,* 10(5), 272-284.

Ryan, R., Hill, S., Lowe, D., Allen, K., Taylor, M. and Mead, C. (2011) 'Notification and support for people exposed to the risk of Creutzfeldt‐Jakob disease (CJD)(or other prion diseases) through medical treatment

(iatrogenically)', *Cochrane Database of Systematic Reviews*, Issue 3 Art. No CD007578available: http://cochranelibrary-wiley.com/doi/10.1002.14651858.CD007578.pub2/full [accessed 20/06/2018].

Schwehr, B. (2018) 'What the revised continuing healthcare framework means for practice', *Community Care*, available: http://www.communitycare.co.uk/2018/03/22/revised-continuing-healthcare-framework-means-practice/ [accessed 05/07/2018].

Simcock, Manthorpe, Tesfamichael, S. and Mead, S. (2018) *Creutzfeldt-jacob disease: guidelines for social workers in England*, London, Department of Health, https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/719058/creutzfeldt-jakob_disease-guidelines-for-social-workers-in-england.pdf (accessed 10/11/2018)

Tricco, A. C., Lillie, E., Zarin, W., O'Brien, K., Colquhoun, H., Kastner, M., Levac, D., Ng, C., Sharpe, J. P. and Wilson, K. (2016) 'A scoping review on the conduct and reporting of scoping reviews', *BMC Medical Research Methodology,* 16(1), 15.

*Wickenden, P. D. (2006) 'Two cases of variant Creutzfeldt-Jakob Disease (vCJD) referred to the Department of Community Mental Health, Aldershot Garrison in 2003', *Journal of the Royal Army Medical Corps,* 152(1), 30-6.